

Clinical Cases.

REPORT OF A CASE OF PROGRESSIVE MUSCULAR ATROPHY WITH BULBAR SYMPTOMS; THE ATROPHY LIMITED TO THE LEFT UPPER EXTREMITY, THE ABDUCTOR INDICIS OF THE RIGHT HAND, AND THE TONGUE, WHICH IS AFFECTED BILATERALLY, PREDOMINANTLY ON THE LEFT SIDE.¹

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I would hardly be justified in presenting to a society devoted to neurology a single case having the usual features of progressive muscular atrophy. On the other hand, the study of such cases as present deviations from the common type, even if such deviation be not great, are frequently of value, if they serve no other purpose than that of exhibiting the non-conformity of disease to our arbitrary standards of classification and description of types of disease.

The patient before you shows peculiarities due to the pathological process having advanced to an extreme degree in certain regions, while its distribution has remained limited; not involving neighboring parts usually affected; the left upper extremity and the tongue being the parts invaded, and these to a marked degree, while the right upper extremity is normal, with the exception of the abductor indicis; the trunk and lower extremities remaining healthy.

His history is as follows A male, æt. 39, a native of Sweden, of occupation a carpenter, married, of temperate habits, and with no history of syphilis or other disease. One year ago, two weeks after his arrival in the United States, observed the first symptom of his present disease, namely, a difficulty in articulation from a slowness of certain movements of the tongue. A month later, weakness in the left thumb and forefinger became troublesome in holding a nail. Weakness of the biceps was observed next, and

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by another month the entire left upper extremity, including the shoulder movements, became perceptibly paretic. A month later, atrophy of the small muscles of the left hand, chiefly the abductor indicis, was detected by the patient. Paresis and atrophy slowly increased, most rapidly in the tongue; deglutition became affected to a slight degree three months after the speech disturbances. Salivation also increased. Fibrillary contractions and the sensations which accompany them were observed by the patient in the upper and lower extremities and in the trunk, but no other sensory disturbances were noticed except that of a deep-seated, stabbing pain in the left biceps, not intense, and a sensation in the tongue which he describes as "a sleepy feeling." Taste has failed during the past year, particularly for delicate tests. He gives no history of other disturbances of the special senses. His mental, alimentary, urinary, and genital functions have remained normal.

Family History.—His father died with dropsy. His mother had some disturbances of speech, lasting for six months, between her fortieth and fiftieth years. She still lives and is eighty years old. He has five sisters and two brothers, all older than himself, who are all alive and without history of disease except one sister who died during childbirth.

The patient was first seen by me at Dr. Seguin's clinic for nervous diseases at the College of Physicians and Surgeons, and was referred to my service at Manhattan Eye and Ear Hospital for electrical treatment.

Examination.—Tests of vision and ocular mobility were kindly made for me by my friend, Dr. J. B. Emerson, at the Manhattan Eye and Ear Hospital.

R. V. Hyperopia $\frac{1}{8}$. Accommodation $\frac{1}{4}$. No insufficiency. L. V. Hyperopia $\frac{1}{10}$.

Pupil normal, iris reacts normally to both light and accommodation. Fundus normal.

Hearing normal.

Facial movements normal except orbicularis oris. Attempts to whistle are imperfect, sucking movements and kissing movements are also imperfect, the labial sounds in speech are weak.

The tongue presents a convoluted appearance of its mucous membrane, due to muscular atrophy, more marked on the left side. He cannot lift the tongue from its bed, nor touch the hard palate with the tip of the tongue, but can protrude it imperfectly and make slow lateral movements. Fibrillary contractions are present to a marked degree.

The left palatal arch is somewhat lower than the right. The throat reflexes are very active. The disturbances in deglutition appear to be due more to defective lingual movements than to abnormal pharyngeal action. The inequality in the palatal arches may possibly be due to the same cause, as it is only apparent in a passive state.

Speech.—The lingual sounds are the most defective, the labials less so. These defects are not so apparent in the utterance of in-

dividual sounds as in continuous speech, and particularly in such words as require sudden changes in the position of the tongue. His wife's name, for example, "Tilla," is difficult for him to articulate. The sound "Th" is quite defective and others which require the approximation of the tip of the tongue to the hard palate. While his speech is not strictly speaking syllabic, there is a slowness of articulation which makes syllabication more distinct than in normal speech; at the same time this slowness is not at all like the slow and slurring speech met with in cerebral cortical degeneration. He can hum tunes as correctly as ever, and his laryngeal functions are normally performed. He thinks, however, that he is not as strong as formerly in shouting in the open air. He has no shortness of breath. Can run as fast and as far as ever, but with more fatigue than formerly. There are no cardiac or pulmonary defects.

Motility, sensibility, and nutrition of the lower extremities are normal, except occasional slight fibrillary contractions. The patellar tendon reflex is active and equal on both sides. The cutaneous reflexes are normal.

Upper Extremities.—The muscles of the entire left upper extremity are of less volume and more flabby than those of the right side. In the left hand, of the abductor indicis but little muscular tissue remains; that which is present abducts the finger alone, but cannot overcome much additional resistance. The thenar group of muscles are flabby and diminished in volume; the dorsal interossei and abductor minimi digiti also show diminished volume and enfeebled power. The flexors and extensors in the forearm, though fairly well preserved, lack the rounded form and the hardness of the corresponding muscles on the right side. The biceps exhibits decided wasting flabbiness and feeble action. The deltoid, the pectorals, and the infraspinatus also show diminished power and volume, particularly the last-named muscle.

The electrical reactions are shown in detail in the accompanying chart.¹ They may be summarized as follows: There was diminished faradic excitability of all the affected muscles, as compared with those of the opposite side; this diminution being relatively greater on direct excitation than through the nerve, even the few remaining fibres of the left abductor indicis responding proportionately to its volume. Slowness of contraction was apparent, particularly in the small muscles of the hand, and to a less degree in the biceps. The abductor indicis was the only muscle of the right side which presented diminution and slowness of contraction. With a reversible electrode, difference in polar action was noticeable in both the right and left abductor indicis, consisting in a reversal of the normal formula of contraction, the anode producing a stronger contraction than the cathode.

With Galvanism.—Quantitatively diminished reactions were found in the affected muscles by direct excitation, while through

¹ Omitted in publication.

the nerves, excepting the left abductor indicis, the reactions were about equal to those of the opposite side when allowance was made for the difference in the volume of the muscles. Qualitative changes, consisting in slowness of contraction and a reversal, complete or partial, of the formula of contraction, were found in both the right and left abductor indicis, on the second interosseous and the ulnar distribution of the thenar group of the left side. The reaction on the left abductor indicis by excitation of the ulnar at the elbow, besides being quantitatively *increased*, gave the unusual result of an anodal closure contraction equal to or greater than the cathodal closure contraction.

The reactions of the biceps to galvanism are interesting, as showing the result of currents of different strength on the formula of contraction in a degenerating muscle.

With 6 M.A. the reaction (by direct excitation) was

K.C.C. > A.C.C.

With 8 M.A.

K.C.C. = A.C.C.

With 10 M.A. < A.C.C.

The anodal contraction was the one which increased in volume, the cathodal being nearly as great with 8 M.A. as with 10 M.A.

The muscles of both sides show increased excitability to mechanical excitation.

Sensibility.—While the patient has been under treatment at the hospital, both Dr. Cramer and myself found diminished sensibility to both the faradic and the galvanic current on the entire left upper extremity, but not elsewhere. At the last examination, December 2d, this could no longer be detected.

In testing sensation with the galvanic current, a considerable difference in resistance was observed between the two sides on the finger tips, which on measurement was found to be 1,000 ohms, showing the importance of working with a milliampère meter in making quantitative tests. If the number of cells required to produce like sensations on the two sides had been taken as a measure, the fallacious conclusion would have been reached that one side was more sensitive than the other.

In the tongue, quantitative diminution, slowness of contraction, and reversal of the formula of contraction were found on direct excitation with both the faradic and the galvanic current.